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# Cerebral Metastasis of Alveolar Soft-Part Sarcoma : Report of a Case of Long Postoperative Survival

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症 例

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## Cerebral Metastasis of Alveolar Soft-Part Sarcoma Report of a Case of Long Postoperative Survival

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In 1952, CHRISTOPHERSON et al<sup>3)</sup> published a new clinicopathological entity "alveolar soft-part sarcoma", which was descriptive of the histological appearance. Although this entity of malignant soft tissue tumor widely has been recognized in recent years, much uncertainty still exists as to the origin, behavior and treatment of this tumor<sup>4)</sup>. In the present communication, we present a case of this tumor, which may be unique in that a long postoperative survival of 11 years 10 months was obtained following total removal of a primary tumor at the left buttock and of a cerebral metastatic tumor.

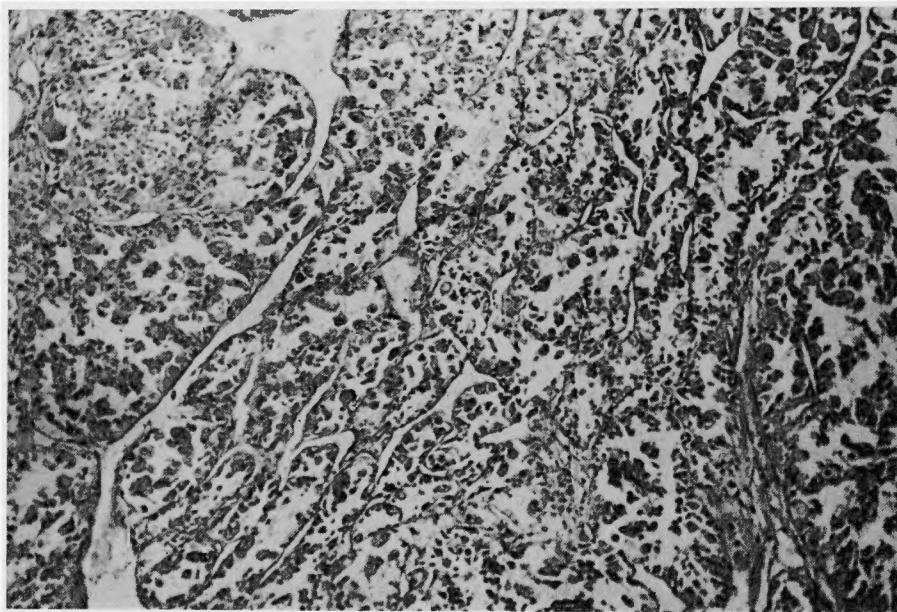
### Case Report

K.H., a 45 year-old woman, was admitted to the 2nd Department of surgery, Gifu University Hospital on February 26, 1958, with complaints of visual disturbance, headache, gait disturbance, sensory disturbance of the extremities and memory disturbance of nine months duration. About 20 years before she fell down on her buttock where she noticed a painful swelling. The swelling had persisted later as a painless lump at the left buttock.

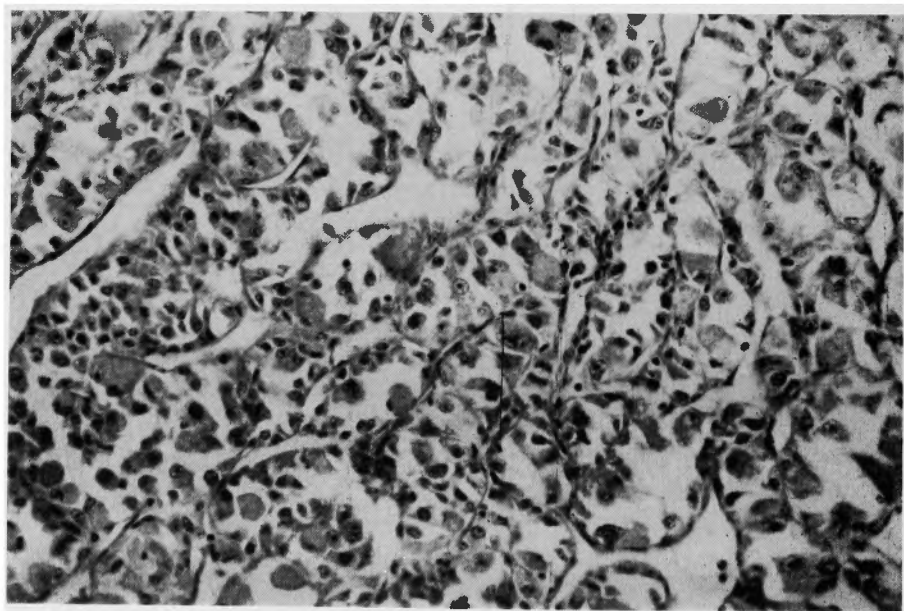
On admission, the patient was emotionally unstable and had memory disturbance. She complained of severe pain in the left parietal region but there was no neck stiffness. Examination of the eyes revealed anisocoria ( $r < 1$ ), absent light reflex on both sides, absent accommodation reflex on the left side, absent corneal reflex on the left side and horizontal nystagmus. Her vision was lost on both sides. Fundoscopically bilateral optic atrophy was noted. On the right side, knee and ankle reflexes were hyperactive. BABINSKI, ROSSOLIMO, OPPENHEIM and CHADDOCK reflexes were positive on the right side. There was a fist-sized painless indurated mass at the left buttock, which was adherent to the gluteal muscle but not to the skin.

Lumbar puncture revealed a clear and colorless CSF under a pressure equivalent to 400 mm of water. Its protein and glucose contents were within normal limits. Chest roentgenogram was not remarkable. Skull roentgenogram demonstrated a thinned dorsum sellae indicating a raised intracranial pressure. Left carotid angiogram revealed that the left anterior cerebral artery was displaced to the right side and the left middle cerebral artery was displaced craniomedially, in anteroposterior view. In lateral view, the left anterior cerebral artery was stretched and the left middle cerebral artery was displaced cranially. An abnormal

vascularity was noted in the left temporal lobe both in arteriogram and in phlebogram. The angiographic diagnosis was a glioblastoma multiforme of the left temporal lobe. Left temporal craniotomy was performed on March 6, 1958. A firm mass was found at a depth of 1 cm beneath the surface of the anterior temporal lobe. Through a cortical incision, a round,



**Fig. 1** Low-power photomicrograph of cerebral lesion showing characteristic pattern of alveolar soft-part sarcoma.



**Fig. 2** High-power photomicrograph of cerebral lesion.

circumscribed, red-purplish and fairly vascular tumor of hen's egg size was found and removed totally. The removed tumor measured approximately  $5 \times 5 \times 6$  cm<sup>3</sup>. It had a dense capsule of connective tissue and white-yellow cut surface, and contained firm tissues in which there was central necrosis and hemorrhage. The outer surface was slightly nodular. Histological study showed an alveolar arrangement of the tumor cells which was consistent with the original description of CHRISTOPHERSON<sup>3)</sup> (Fig. 1. 2.).

It was then postulated that the painless indurated mass at the left buttock might be the primary tumor. It was removed operatively on May 21, 1958, and the same histological finding as above was obtained.

Postoperatively, the patient made an uneventful recovery. The visual impairment persisted, but the other complaints were improved. She was discharged on June. 13, 1958. Follow-up over the next 11 years indicated that she was doing well, with no symptoms except for visual deficit. On June, 1969, she came to complain of headache, convulsion, vomiting, and speech disturbance. Following gradual exacerbation of these symptoms she died on January 11, 1970. No local recurrence of tumor at the left buttock was noted by her home doctor. Although autopsy was not performed, it is highly probable that she died of recurrence of brain metastasis. She survived 11 years and 10 months following removals of the primary tumor at the buttock and the metastatic tumor in the brain.

### Discussion

In 1966, LIBERMAN et al<sup>4)</sup>. analyzed their 53 cases of alveolar soft-part sarcoma, and pointed out that the location, lateralization, particular age distribution, female predilection and indolent but inexorable course separated alveolar soft-part sarcoma biologically from other soft-tissue tumors. Forty-six cases of them were followed long enough to permit collection of survival data by actuarial methods: 82.8% in two years, 59% in five years and 47.1% in ten years. Their two patients observed the longest died of disseminated sarcoma in the 20th year, so there were no lifetime cures.

The striking histologic feature of this tumor is pseudoalveolar or organoid arrangement of cells in relation to numerous delicate endothelial lined vascular channels and septa. ROSENBAUM et al<sup>5)</sup>. indicated that the unusual juxtaposition of the tumor cells to the endothelium seems responsible for the highly vascular appearance of the lesion on angiography, the frequency of hematogenous spread and the grave, although delayed, prognosis. Even though its course seems slow giving surprisingly few symptoms even with extensive metastases, it seems to be a very difficult disease to cure. According to LIBERMAN et al<sup>4)</sup>. 28 of their 53 cases of the neoplasm had metastases proved pathologically or by x-ray examinations, or both. The most frequent sites of metastases were lung (12%), bone (19%) and brain (15%). CHRISTOPHERSON et al.<sup>3)</sup> reported 2 cases of brain metastases in their 12 cases of the neoplasm. Neither authors performed craniotomy for brain metastasis.

ASVALL et al.<sup>1)</sup> indicated that the primary surgery should be radical but, by contrast, LIBERMAN et al<sup>4)</sup>. concluded that extensive surgery proved of no great value beyond palliation.

In the present case, both the primary tumor and the metastatic brain tumor were fairly well circumscribed and their extirpation was considered to be practically complete, therefore no additional radiation or chemotherapy was performed. It may be that the long postoperative survival in our case is related to the fact that both the primary and the metastatic tumors were fairly sharply bordered with normal tissues and could be removed totally. If so, the clinical course of our case may lend support to the opinion of ASVALL et al<sup>1)</sup>, that radical removal, whenever possible, is the treatment of choice. ROSENBAUM et al<sup>5)</sup>, reported two cases of brain metastasis of the neoplasm, one patient being irradiated and the other operated upon, and the latter did well postoperatively. They did not describe survival period of the patient. As for conservative treatments, radiosensitivity of the neoplasm has been reported to be low<sup>6)</sup> or moderate<sup>1)</sup>, while favorable effects of chemotherapy have been reported.<sup>1,7)</sup>

A comment will be made on significance of trauma as an etiological factor of this neoplasm. CHRISTOPHERSON et al.<sup>3)</sup> considered that a history of mild trauma, which often had been found in their cases, was nothing more than calling attention to the mass, while BATESON<sup>2)</sup>, in presenting a case of the neoplasm arising in the right thigh following a kick thereupon, stated that the tumor of his case possibly had arisen or had been aggravated by the trauma. In the present case there was a history of contusion on the left buttock, the same site as the primary tumor, and this fact may be in accordance with BATESON's<sup>2)</sup> opinion, although no definite conclusion can be drawn in this respect.

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## 和文抄録

Alveolar Soft-part Sarcoma 脳転移  
——術後長期生存の1例——

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大橋広文・坂田一記・上田茂夫・坂本武嗣・三輪 勝

左臀部に原発し、左側頭葉に転移を来した alveolar soft-part sarcoma の45才女性症例において原発及び転移腫瘍の摘出術を行い、患者は術後11年10カ月生存した。

脳転移摘出後長期生存する例は珍しい、本症例が

長期生存し得た理由は、原発及び転移腫瘍がいずれも限局しており全摘出が可能であったためと思われる。

本症例においては原発腫瘍が発生した部位に外傷の既往が認められ、外傷と腫瘍発生との間の因果関係の存在が想定された。